Limb amputation and other disability desires as a medical condition

Brugger, Peter ; Christen, Markus ; Jellestad, Lena ; Hänggi, Jürgen

Abstract: Some people have a profound dissatisfaction with what is considered an able-bodied state by most others. These individuals desire to be disabled, by conventional standards. In this Review, we integrate research findings about the desire for a major limb amputation or paralysis (xenomelia). Neuropsychological and neuroimaging explorations of xenomelia show functional and structural abnormalities in predominantly right hemisphere cortical circuits of higher-order bodily representation, including affective and sexual aspects of corporeal awareness. These neural underpinnings of xenomelia do not necessarily imply a neurological cause, and a full understanding of the condition requires consideration of the interface between neural and social contributions to the bodily self and the concept of disability. Irrespective of cause, disability desires are accompanied by a disabling bodily dysphoria, in many respects similar to gender dysphoria, and we suggest that they should be considered a mental disorder.

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The desire for limb amputation or paralysis: a medical condition?

Peter Brugger[1,2,3,*] Markus Christen[4], Lena Jellestad[5]
Jürgen Hänggi [6]

[1] Neuropsychology Unit, Department of Neurology, University Hospital Zurich, Zurich, Switzerland

[2] Center for Integrative Human Physiology (ZIHP), University of Zurich, Zurich, Switzerland

[3] Neuroscience Center Zurich, University of Zurich and ETH Zurich, Zurich, Switzerland

[4] University Research Priority Program Ethics, University of Zurich, Zurich, Switzerland

[5] Department of Psychiatry, University Hospital Zurich, Zurich, Switzerland

[6] Division of Neuropsychology, Department of Psychology, University of Zurich, Zurich, Switzerland

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***Correspondence:**

Dr. Peter Brugger
Neuropsychology Unit
Department of Neurology
Frauenklinikstrasse 26
CH-8091 Zürich, Switzerland
Phone: ++41 44 255 5570
Peter.brugger@usz.ch

Summary

There are persons who suffer from a profound dissatisfaction with what is considered an able-bodied state by most others. They desire to be "dis-abled" by conventional standards. We integrate recent research findings on the desire for a major limb amputation or paralysis ("xenomelia"). Neuropsychological and neuroimaging explorations of xenomelia revealed functional and structural abnormalities in predominantly right hemisphere cortical circuits of higher-order bodily representation, including affective and sexual aspects of corporeal awareness. These neural underpinnings of xenomelia do not necessarily imply a neurological etiology, and a full understanding of the condition requires consideration of the interface between neural and social contributions to the bodily self and the concept of (dis)ability. Irrespective of etiology, disability desires are accompanied by a disabling bodily dysphoria, in many respects similar to gender dysphoria, and deserve to be considered a mental disorder.

Introduction

This review integrates recent research findings concerning a condition that is not yet recognized as a mental disorder, which is still in search of a generally accepted medical label and whose conceptual basis is still "under construction". The condition involves a person's dissatisfaction with an able-bodied state. It is referred to as "body integrity identity disorder" (BIID) in the psychiatric literature, "xenomelia" in neurologically oriented studies, and "transability" ("transableism") in approaches to disability from a primarily sociological perspective (table 1, appendix and figure 1, appendix, for the terminology). In this review we use the term "disability desires" to remain descriptive and avoid controversial concepts of identity and too narrow notions of focal brain damage. Although deafness, blindness or the status of a eunuch can be the target of disability desires, the focus of our review is on the desire for limb amputation or paraplegia. These forms of profound bodily modification are the most frequently reported in case reports and surveys. Also, empirical data beyond those collected in questionnaire studies are available only from persons who wish to get rid of one or more of their limbs.

Core features of disability desires

Persons with a disability desire describe a profound mismatch between their actual and their desired body, with respect to its shape or functionality. A frequently used word in their complaints is that of "overcompleteness". In the most common variant, the desire for amputation, persons suffer from having four limbs, because their felt identity is that of an amputee, and their "inner body" does not match their physical appearance (panel 1, for typical first-hand descriptions). In a case of desired hearing loss, normal hearing was experienced as a source of stress and fear in the form of hyperacusis and misophonia,[11] and the longing for blindness may be described as a suffering from regular light conditions . Not uncommon is the explicit statement that the desire for a certain disability is not stronger than the desire to compensate for it by use of prosthetics, such as crutches, a wheelchair or hearing-aids. In fact, "pretending" to already have the desired disability and to be dependent on the respective aids is an important, almost ubiquitous aspect of the condition (panels 1 and 2). There is agreement about an onset of disability desires in childhood or early adolescence, "between the ages 5 and 15".[24, p.28]

Prevalence and demographics

The overall prevalence of disability desires in the general population is unknown. It is higher in Western cultures promoting societal individualism,[25]but single cases have been reported in Asian countries.[26] On the Internet there is a growing number of "virtual communities" devoted to disability desires, but to derive prevalence estimates from this fact is barely possible.

An overwhelming preponderance of the male gender is undisputed. This is more pronounced for the desire for limb amputation than for paraplegia,[27] and possibly even lower for disability desires targeting seeing and hearing.[28] The prevalence of non-heterosexuality among persons with disability desires is controversial (panel 2, table 1); in the largest surveys to date it was found to be 38% (n=52),[1] 33% (n=72), [17] and 44·5% (n=54).[8] This is well above the prevalence in the population at large (approx. 2%),[29] but the reasons for the association between disability desires and sexual orientation are unknown.

Proposed diagnostic criteria and differential diagnosis

Disability desires are not currently listed in DSM-5 or ICD-10. Panel 3 displays the diagnostic criteria proposed by advocates of its future classification as a mental disorder under the label "Body Integrity Identity Disorder".[16] In brief, the desire to become disabled must be

longstanding and intense. It produces persistent discomfort, and attempts to reach a disabled state may lead to self-inflicted harm.[4,30,31] Onset of the disorder is required to be before adulthood.
The desire for major body modifications must not be rooted in a psychotic disorder, where self-amputations and mutilation fantasies are occasionally observed. Also acute brain damage should be excluded as cerebral lesions can be accompanied by an estrangement from one's own limbs. Frequently discussed in the context of the desire for limb amputation are somatoparaphrenia (i.e. the denial of ownership over functionally impaired limbs) and misoplegia (i.e. the hatred of own body parts). [32] Although these and other neurological syndromes are superficially linked to disability desires (table 2, appendix), none of them captures their essence, i.e. the non-delusional fantasy, in the absence of sensory misperceptions, that some body modification would enable a person to reach her "true identity". All these syndromes have a sudden onset (most typically they follow a cerebro-vascular incident) whereas disability desires are a *developmental* disorder in the experienced unity of body and self. A one-to-one-correspondence in phenomenology must not, therefore, be expected. Body dysmorphic disorder is sometimes confounded with disability desires. [3,33,34] The two conditions may share a preoccupation with a specific part of one's body, but body dysmorphic disorder is typically concerned with the visual aspects of that part, which is never the case in disability desires. Also, the concerns in body dysmorphic disorders usually focus on facial parts, not on major limbs.

There are close conceptual links between the desire for healthy limb amputation and gender dysphoria. [14,25] In fact, the term "body integrity identity disorder" (BIID) was modelled after that of "gender identity disorder" (GID) by simply substituting "gender" by "body integrity". [1] Table 2 lists some of the commonalities between the conditions. They comprise an early onset, a predominance of the male sex and the frequent simulation of the desired state. An elevated prevalence of non-righthandedness points to abnormalities at early neurodevelopmental stages, but the relevant neural structures have not yet been identified. Proposed cerebral loci and circuits appear to increase with an increasing number of studies both for GID[38] and for disability desires (see section on xenomelia, below).

Before being conceptualized as an identity disorder, the desire for amputation was considered a paraphilia. "Apotemnophilia" [4] denotes the sexual arousal by the fantasy of being an amputee and is, more often than not, accompanied by "acrotomophilia",[39] the sexual arousal by others' disability (notably an amputation). However, those sufferers lobbying for an inclusion of amputation desires in the DSM argued that they would not have a paraphilic origin at all. Bridy noted that "Apotemnophiles and their advocates today are at pains to emphasize that apotemnophilia is a broad-based identity disorder and not one that is narrowly sexually determined". [40, p.149] With the proposal that BIID, like GID, would be an identity disorder [1] disability desires were released from too narrow a focus on sexuality. This happened, however, at the price of downplaying sexual aspects. First, in his seminal study with 52 persons with disability desires,[1] emphasized that only 15% of the participants indicated sexual arousal as the *primary* motivation for their desire. It was later pointed out[15,33] that another 52% indicated sexual arousal at least as their *secondary* motivation. Also, 45 of First's 52 participants reported being sexually attracted to amputees – thus 87% had paraphilic desires on top of an affliction of "purely bodily identity"! More recent surveys produce similar figures; one study recruited 54 persons with a disability desire on the Internet and found the percentage of persons reporting "specific sexual desires" when imagining a disabled

state was 90·7%.[8] Even in the desire for hearing loss, the power of the "ear-otic" dimension was emphasized.[28,p.366] In a planned study of the brain morphometric correlates of disability desires we intended to recruit participants with an exclusively asexually based amputation desire, but the recruitment via the Internet revealed comparable ratings of the strengths of sexually neutral versus sexually arousing components of an individual's desires.[41] Thus, as far as disability desires are concerned, sexual undertones seem to be always part of the bigger picture of corporeal identity. Just how large this part will be, or will be reported to be, in an individual case, may depend on the criteria that define disability desires. Because of the inseparability of sexuality and identity, we suggest that the first part of point D in the proposed diagnostic criteria (panel 3) should be omitted.

The two latest efforts in the terminology of disability desires (figure 1, appendix) tried to circumvent allusion both to the paraphilias and the concept of identity. "Transableism"[42] is a sociological construct which attempts to demedicalize disability desires and views them as a healthy person's challenge to the stigma of disability as created by social norms. "Xenomelia"[43] may be positioned at the opposite end of a continuum from social to biological determinants of a disease. It conceives of disability desires as resulting from developmental brain damage. There may be limitations of the concept (table 1, appendix), but work on xenomelia has produced a large body of empirical data.

Xenomelia: a neurological perspective on disability desires

“Xenomelia” (from Greek хечф (xeno) = foreign and μελοσ (melos) = limb) denotes estrangement from one's own limbs. The term was introduced by McGeoch et al.[43] to counter previous, theory-laden terms, notably apotemnophilia and BIID. The authors suggested that disability desires represent a neurological disorder, specifically a focal syndrome of the right parietal lobe. Clinical neurology identifies many syndromes of various misperceptions of body parts after damage to this site of the brain, ranging from a total neglect of the left side of the body to illusory reduplications, the loss of agency and ownership up to an active aversion or hatred of left-sided limbs (table 2, appendix). [44]

Several clinical observations support a neurological origin of disability desires concerning arms or legs. First, the type of a desired disability is well circumscribed and developmentally stable. A person longing for a bilateral leg amputation usually abhors the idea of becoming paraplegic (and vice versa). Furthermore, left-sided limbs are far more frequently the target of an amputation desire than right-sided limbs (table 1). This is in line with the leading role of the right cerebral hemisphere in the representation of the bodily self and its disorders[44,45]. The right parietal cortex also codes for left- and right-sided limbs, which explains why exceptional switches of an amputation desire to the contralateral limb[46] do not invalidate the neurological hypothesis. Finally, there is commonly a demarcation line precisely indicating the site of a desired amputation.

Some behavioral experiments have exploited this clear separation between what is considered own body territory and what is felt to be foreign. They showed that tactile stimulation of the latter regions are accompanied by an elevated autonomous response[47,48] and a priorization in the judgment of temporal order. [49] This pattern is indicative of a *hyper*attention towards incoming stimuli. Other experiments have compared the behavior of persons with xenomelia with that of patients with somatoparaphrenia. In both conditions Romano et al. found a reduced anticipation of pain, which was specific to the disowned or non-accepted limb.[48,50] Similarities (and differences) between the two conditions were also shown in an illusion paradigm that requires the integration of vision, touch and proprioception. [51,52] Caloric

vestibular stimulation, known to transiently abolish somatoparaphrenic delusions,[53] did not affect the desire for amputation as assessed by a questionnaire. [54] Taken together, somatoparaphrenia and xenomelia may differ along explicit variables, but share commonalities in attentional and autonomous nervous system functions. Using a conceptually distinct approach, a study of emotional processing in persons with xenomelia reported a selectively reduced disgust sensitivity in response to pictures of body violations, which was interpreted as compatible with insular dysfunction.[55] The report highlights the importance of considering aspects of body representation beyond those of "body schema" in the narrow sense.

Neuroimaging studies have addressed brain-behavior relationships in xenomelia more directly. A first experiment examined the neural response to tactile stimulation in four individuals with a desire for a leg amputation and four healthy control persons.[43] Stimuli were taps delivered to the feet and anterior thighs, well above an individual's demarcation line (and to mirrored locations on the healthy leg and on both control subjects' legs). Magnetoencephalographic signals to taps on the non-accepted compared to the accepted legs were reduced in the xenomelia persons' right superior parietal lobe (SPL), a reduction seen also in the comparison with healthy persons' legs (Fig.1A and B). Crucially, the diminished response of the right SPL was independent of whether the amputation desire was targeting the left or the right leg, supporting the known bilateral body representations of specifically the right hemisphere.[56] A functional magnetic resonance imaging (fMRI) experiment with five persons who desired a leg amputation (3 of the right leg, 2 of the left), found an increased neural response to tactile stimulation on the lower leg compared to corresponding stimulations in healthy control participants.[57] Areas of increased activity were found in a widespread network comprising parieto-frontal and occipito-temporal cortex bilaterally (pronounced in the right hemisphere) and the right insula (Fig.1D). Interestingly, this enhanced response was independent of whether the affected or the non-affected leg was stimulated. However, the difference in the response between the two legs interacted with group membership: for xenomelia participants, but not for controls, touch on the non-accepted leg triggered a smaller premotor cortex response than that on the accepted legs (Fig.1C). Together, these two findings indicate that xenomelia is associated with a generally higher responsivity to tactile stimulation, which might be due to an elevated attention to tactile stimuli. Ownership feelings may not primarily depend on activity in parieto-insular networks (as one might have expected from research on somatoparaphrenia,[58,59]) but also on premotor cortex, whose role for mediating ownership over body parts had been demonstrated before.[60]

Structural brain correlates of xenomelia were investigated in 13 men suffering from the desire for leg amputation (8 for the left leg, 2 for the right, 3 for both legs).[61,62] Surface-based morphometry based on T1-weighted MRI revealed, compared to 13 matched control persons, a reduced cortical thickness of the superior parietal lobule, in close topic correspondence to the functional impairments reported earlier and likewise confined to the right hemisphere[43]. A reduced cortical surface area was described for the right inferior parietal lobule, primary and secondary somatosensory cortex and the right anterior insula[61] (Fig. 3A, left). The strength of an individual's amputation desire was inversely related to the cortical surface area in the right inferior parietal cluster (Fig. 3A, right). Shape analyses[62] uncovered tissue displacements (local thinning or thickening) in several subcortical structures bilaterally and specifically in subregions known to represent the body in a somatotopic manner or project to motor cortical areas (Fig. 3B–D).

Behavioural and neuroimaging findings in persons with xenomelia have refined our knowledge about the condition. Its neural correlates involve a network of cortical and

subcortical regions, comprising three key regions; the parietal lobes as the classical site of corporeal awareness,[44]the insula, known for its crucial role in the integration of body and mind[63] and the premotor cortex, previously implicated in the formation of the unity of body and self.[60,64] This network is strongly lateralized to the right hemisphere, which is compatible with the preference for left-sided body parts as targets for amputation desires and with an elevated incidence of non-righthandedness in persons with disability desires (tables 1 and 2).

However, the shortcomings of a conceptualization of disability desires too narrowly based on physiology and neuropsychology are evident. On the one hand, all evidence for a focal dysfunction in higher-order sensorimotor circuits for limb representation has been obtained in experiments with persons with xenomelia in the narrow sense. The present-day neurological account of disability desires may thus be valid for the desires for limb amputation or paralysis, but not necessarily for the desire to become blind, deaf or genderless. Moreover, like the traditional concept of "body integrity", xenomelia falls short of capturing all aspects of disability desires, notably their sexual dimension. The notion of disability desires as a parietal lobe syndrome has once more exposed the gap between those adhering to an individualistic view of embodiment and those focusing on sociological notions of a body-in-the-world. Research on disability desires could diminish this great divide between brain-based and mind-based world views by integrating neurological and sociological work on bodily representations.

Towards a social neuroscience of disability desires

To be human means to be a biosocial creature that requires description in both biological and sociological terms. A full understanding of disability desires will require consideration of causative forces beyond gray and white matter and consider social matters as well. Paul Schilder's visionary concept of a neural representation of the human body as a medium between self and others let him recognize that "body-image is a social phenomenon".[65,p.217] He thus anticipated currently popular views of social neuroscience, according to which the ways we feel and experience our own body should be considered avenues to an understanding of how we communicate with conspecifics who, for the most part, have similar bodies.

"Body image" represents our corporeal awareness on the most abstract level; the one which allows us to evaluate our own appearance against esthetic standards and cultural norms.[66] This level of body representation is at the same time the most difficult to pinpoint in terms of cerebral localization. However, work over the past two decades has demonstrated interactions between self and others at much lower levels of perceptual-motor integration. In fact, the neural prerequisits of the human mirror system reside in premotor and somatosensory cortices.[67] An experimental report about neonates' spontaneous imitation behaviour[68] has been supported by electrophysiological recordings in very young infants,[69] which showed that visually observed hand and foot actions are automatically matched in a somatotopic way to the observer's own somatosensory and motor cortices. Such hierarchically and developmentally early processing of other persons' body shape and movements is important for an understanding of some xenomelia-associated behaviors. Approximately half of persons with xenomelia claim that their desire for amputation was triggered by the sight of an amputee.[19] While such introspective reports are reminiscent of the concept of maternal impression[70] and may be considered secondary rationalizations, a neuropsychological basis should not be dismissed on *a priori* grounds. We have speculated[61] that a hyperempathic response might predispose individuals to integrate observed bodily defects into their own body schema. This suggestion was based on the phenomenon of "mirror-touch synaesthesia", observed in a minority of healthy persons and indicating the feeling of touch on visual observation of others being touched.[71] This form of synaesthesia is linked to psychological

empathy[72] and is especially frequent in amputees.[73] In those rare cases of phantom limb in persons with limb aplasia, a trigger function of the visual observation of others' limb movements has been documented.[74] The conceptualization of xenomelia as a mirror image of aplasic phantom limbs[75] is perhaps more than a metaphor; if in the latter case, seeing a limb in motion can elicit feeling a corresponding own limb, in xenomelia, observing *the absence* of another person's limb could unmask a congenital underrepresentation of the own limb – in the words of Robert Smith, the sight of an amputee "seems to awaken an internal identity that had previously been unrecognized". [76,p.73]

The undeniable paraphilic component of the desire for amputation[15] may find an explanation in the architecture of low-level body representations. The adjacency of the foot/leg representation and that of the genital organs in the postcentral gyrus may be more than a coincidence when it comes to explain the four-fold incidence of legs over arms in amputation desires and the stronger erotic connotations of disability desires with *leg* compared to arm amputees.[77] This homuncular vicinity is probably determined during foetal life, induced by the frequency of genital self-touch with the feet due to the typical position of the unborn child.[78] It is the physiological explanation for the fact that touch to the face, but never to the genital region, can elicit phantom sensations in hand amputees, whereas the converse is true for foot amputees. [79] There is a large body of empirical work on the specificity of these "referred sensations". [80] Furthermore, the insula as a hub for the integration of body and mind [63] and, more specifically, of somatosensation and sexual arousal [81] is adjacent to the secondary somatosensory cortex for leg representation. This might explain its repeated inclusion among the hot spots of functional [47] and structural [61] cerebral abnormalities in persons with xenomelia. De Preester[15] localized a "sexual schema" in conceptual proximity to an insular-parieto-frontal system that merges somatosensation, erotic arousal and visual observation of conspecifics. Specifically, she considered conceivable that the observation of an amputee during early development could lead to a "dramatic explication of the sexual schema into the body image".[15,p.183] Clearly, prospective research is needed to substantiate or refute such proposals. What is a fact is that paraphilic components are largely neglected by both advocates of a brain-based and those of a mind-based approach to disability desires. This should be changed in future empirical research (panel 4).

The thoughts presented in this section are speculations. They may inform those who work on the cognitive neuroscience of bodily self-consciousness as it relates to aspects of social life.[45,83] They are, however, neither of direct help to the persons suffering from disability desires nor to the clinicians who aim at treating them. The question arises: do we need to treat disability desires at all? Are they pathological desires or do they merely reflect a normal urge of a minority for "a continual testing to discover what could be incorporated in the body"?[65,p.217]

Disability Desires: divergence or disorder?

Detailed diagnostic criteria for disability desires have been catalogued by the American psychiatrist First who coined the term "BIID" and who is confident that the condition "will eventually be included in a future edition of the DSM". [9,p.853] This opinion is shared by the British surgeon Smith who had operated on several persons with an amputation desire[76] and proposed similar criteria before.[24] In stark opposition to this attempt to provide disability desires an official medical status are those who consider them "a new way to be mad",[84] "a contemporary frame for psychological suffering"[85,p.609] or "the first psychiatric disease 'manufactured' in cyberspace".[86,p.340] They argue that the Internet is a platform that popularises "transient" mental illnesses,[87] whose particular symptom constellation is in steady flux. This constellation is shaped by a continuous exchange between sufferers who share their

experiences in organized chats and the policy makers who compile and classify the reported symptoms.

Both DSM and ICD are endeavours in taxonomy that, far from categorizing illnesses in an objective and neutral way, actively create, shape and eliminate certain conditions. A case in point is homosexuality, a "sociopathic personality disturbance" in DSM-I, a "sexual deviation" in DSM-II, and no longer any disorder from DSM-III onwards.[88] People with disability desires are increasingly lobbying that their condition should be listed, partly to promote the availability of surgical treatment. Given the massive distress accompanying disability desires one might wonder whether BIID would not better be read as BD, for "body dysphoria" (panel 4). Those who feel an urge to use a wheelchair or crutches or to wear special lenses that prevent light penetrating their eyes could do so, but their behaviour would not be considered pathological unless accompanied by considerable distress.

Another question is whether conformance to disability desires can ever be ethically justified? This discussion has unfolded in connection with elective amputations offered to sufferers from xenomelia in some non-Western countries. The framework of bioethical principlism[89] indicates that the controversy is mainly about empirical issues, for instance whether the conditions for claiming respect for autonomy are fulfilled in patients with disability desires or whether there is sufficient evidence for therapeutic success of elective amputations (table 3). Successful psychotherapy of disability desires is rarely reported[20] and probably reduces associated symptoms like depression, but not disability desires *per se*.[99] In contrast, reports about amputations in the case of xenomelia describe an immediate and lasting alleviation of chronic suffering.[1,21,100] (ref. 22 for an exception). The largest case series of surgically remedied disability desires comprises 21 persons, 18 having experienced a major amputation during the past 1 to 16 years.[21] In all cases quality of life was rated to be substantially increased, and no new disability desire emerged post-surgery. Furthermore, evidence shows that competence for autonomous decision making cannot be denied for persons with xenomelia.[76] In summary, there is considerable support for the view that elective amputations can be ethically justified in some cases, even if long-term effects of the intervention still need to be assessed. What is needed is a consensus paper authored by bioethicists, psychiatrists, and surgeons. Some authors specifically ask for regulations that guarantee conforming surgeons' protection from legal sanctions.[76]

Conclusions

Disability desires represent a person's dissatisfaction, since early childhood, with what most people consider an able-bodied state. This dissatisfaction can be so disabling as to justify conceptualization of the condition as a mental disorder. In the case of desired limb amputation there is evidence for an altered cerebral architecture accompanying disability desires. Yet, biological mediation does not imply biological etiology.[101] Bodily self-consciousness is subject to powerful socio-cultural influences, which does also not imply that disability desires have primarily societal roots.[83] Future research should explore the intersections between neural and psychological levels of analysis and promote an ethnologically, sociologically and neuropsychologically informed perspective on disability desires. With respect to ethical concerns about elective amputations, detractors' arguments yield to strong counterargument. However, whether cutting the body will ever cure the mind [102] will remain debatable in the discourse about disability desires and related conditions of bodily dysphoria.

Search strategy and selection criteria

This review is based on material from the authors' personal files and the references in the single articles. A combination of the search terms "body integrity", "identity", "disability",

"amputation", "paraphilia", "xenomelia" and the terms listed in Table 1, appendix, produced more items with the aid of PubMed, the Web-of-Science and Scopus (last accessed May 17, 2016). Selection of the references finally included was based on an article's quality and originality and its usefulness to aid argumentation. Articles in English, French, Italian, Dutch and German were considered.

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Author contributions
All authors contributed to the compilation and selection of work included in this review. PB drafted a first version of the article, and all authors contributed to editing the text and preparing display items. All authors have approved the final version.

Declaration of interests
We declare no competing interests.

References

1. First MB. Desire for amputation of a limb: paraphilia, psychosis, or a new type of identity disorder. *Psychol Med* 2005; **35**: 919–28.
2. Swindell M, Lawrence JS. Body integrity identity disorder: an overview. In: Stirn A, Thiel A, Oddo S, eds. Body integrity identity disorder: Psychological, neurobiological, ethical and legal aspects. Lengerich, Germany: Pabst, 2009: 11–9.
3. Kasten E. Body Integrity Identity Disorder (BIID): Befragung von Betroffenen und Erklärungsansätze. *Fortschr Neurol-Psychiatrie* 2009; **77**: 16–24 (in German).
4. Money J, Jobaris R, Furth G. Apotemnophilia: two cases of self-demand amputation as a paraphilia. *J Sex Res* 1977; **13**: 115–25.
5. Nelson [pseudonym]. Living a life with BIID. In: Stirn A, Thiel A, Oddo S, eds. Body integrity identity disorder: Psychological, neurobiological, ethical and legal aspects. Lengerich, Germany: Pabst, 2009: 82–7.
6. Becker A. Body integrity identity disorder (BIID) and me. In: Stirn A, Thiel A, Oddo S, eds. Body integrity identity disorder: Psychological, neurobiological, ethical and legal aspects. Lengerich, Germany: Pabst, 2009: 103–6.
7. O'Connor S. My life with BIID. In: Stirn A, Thiel A, Oddo S, eds. Body integrity identity disorder: Psychological, neurobiological, ethical and legal aspects. Lengerich, Germany: Pabst, 2009: 88–93.
8. Blom RM, Hennekam RC and Denys D. Body integrity identity disorder. *Plos One* 2012; **7**. Article Number: e34702.
9. Johnson TW, Wassersug RJ, Roberts LF, Brett Sutherland M, First MB. Desire for castration is not a body integrity identity disorder (BIID): a response. *J Sex Med* 2010; *7*: 853–5.
10. Campbell FK. Contours of ableism. The production of disability and abledness. Basingstoke: Macmillan, 2009.
11. Veale D. A compelling desire for deafness. *J Deaf Stud Deaf Educ* 2006; **11**: 369–72.
12. Johnson TW, Irwig MS. The hidden world of self-castration and testicular self-injury. *Nature Rev Urology* 2014; **11**: 297–300.
13. Lawrence AA. Desire for castration is not a body integrity identity disorder (BIID): comments on Johnson et al. (2007), Brett et al. (2007), and Roberts et al. (2008). *J Sex Med* 2010; **7**: 852–53.
14. Lawrence AA. Clinical and theoretical parallels between desire for limb amputation and gender identity disorder. *Arch Sex Behav* 2006; **35**: 263-78.
15. De Preester H. Merleau-Ponty's sexual schema and the sexual component of body integrity identity disorder. *Med Health Care and Philos* 2003; **16**: 171–84.
16. First MB, Fisher CE. Body integrity identity disorder: the persistent desire to acquire a physical disability. *Psychopathology* 2012; **45**: 3–14.
17. Johnson AJ, Liew S-L, Aziz-Zadeh L. Demographics, learning and imitation, and body schema in body integrity identity disorder. *Indiana Univ Undergrad J Cog Sci* 2011; **6**: 8–15.
18. Blanke O, Morgenthaler FD, Brugger P, Overney LS. Preliminary evidence for a fronto-parietal dysfunction in able-bodied participants with a desire for limb amputation. *J Neuropsychol* 2009; **3**: 181–200.
19. Bou Khalil R, Richa S. Apotemnophilia or body integrity identity disorder: a case report review. *Int J Lower Extr Wounds* 2012; **11**: 313–19.
20. Thiel A, Ehni FJ, Oddo S, Stirn A. Body integrity identity disorder - first success in long-term psychotherapy. *Psychiat Praxis* 2011; **38**: 256–8 (in German).

21. Noll S, Kasten E. Body integrity identity disorder: how satisfied are successful wannabes? *Psychol Behav Sci* 2014; **3**: 222–32.
22. Sorene ED, Heras-Palou C, Burke FD. Self-amputation of a healthy hand: A case of body integrity identity disorder. *J Hand Surg Br* 2006; **31**: 593–5.
23. Oddo S, Möller J, Skoruppa S, Stirn A. Psychological features of BIID: Personality traits, interpersonal aspects, coping mechanisms regarding stress and conflicts, body perception. *Fortschr Neurol Psychiatr* 2014; **82**: 250–60 (in German).
24. Smith R. Amputee identity disorder and related paraphilias. *Psychiatry* 2004; **3**: 27–30.
25. Lawrence AA. Parallels between gender identity disorder and body integrity identity disorder: a review and update. In: Stirn A, Thiel A, Oddo S, eds. Body integrity identity disorder: Psychological, neurobiological, ethical and legal aspects. Lengerich, Germany: Pabst, 2009: 154–72.
26. Blom RM, Vulink NC, van der Wal SJ et al. Body integrity identity disorder crosses culture: case reports in the Japanese and Chinese literature. *Neuropsychiatric Dis Treat* 2016; **12**: 1419-23.
27. Giummarra MJ, Bradshaw JL, Hilti LM, Nicholls ME, Brugger P. Paralyzed by desire: a new type of body integrity identity disorder. *Cog Behav Neurol* 2012; **25**: 34–41.
28. Harmon K. Hearing aid lovers, pretenders, and deaf wannabes: the fetishizing of hearing. In: McRuer R, Mollow A, eds. Sex and disability. Durham and London: Duke University Press, 2012: 355–72.
29. Ward BW, Dahlhamper JM, Galinsky AM, Joestl SS. Sexual orientation and health among U.S. adults: national health interview survey, 2013. *Nat Health Stat Rep* 2014; **77**: 1–10.
30. Beresford TP. The dynamics of aggression in an amputee. A case report. *Gen Hosp Psychiatry* 1980; **2**: 219–25.
31. Clervoy P, Vautier V, Naudin J. Body identity integrity disorder: clinical questioning and ethical issues. *Ann Méd-Psychol* 2009; **167**: 251–5 (in French).
32. Giummarra MJ, Bradshaw JL, Nicholls ME, Hilti LM, Brugger P. Body integrity identity disorder: deranged body processing, right fronto-parietal dysfunction, and phenomenological experience of body incongruity. *Neuropsychol Rev* 2011; **21**: 320–33.
33. Bayne T, Levy N. Amputees by choice: body integrity identity disorder and the ethics of amputation. *J Applied Philos* 2005; **22**: 75–86.
34. Ryan CJ, Shaw T. BIID is nor BDD – comment on Kwok-Kwan Chan et al. *J Plastic Reconstr Aesthetic Surg* 2011; **64**: 8–9.
35. Veale JF, Clarke DE, Lomax TC. Biological and psychosocial correlates of adult gender-variant identities: a review. *Pers Indiv Diff* 2010; **48**: 357–66.
36. Lawrence AA. Sexuality before and after male-to-female sex reassignment surgery. *Arch Sex Behav* 2005; **34**: 147–66.
37. Berger BD, Lehrmann JA, Larson G, Alverno L, Tsao CI. Nonpsychotic, nonparaphilic self-amputation and the internet *Compr Psychiatry* 2005; **46**: 380–3.
38. Smith ES, Junger J, Derntl B, Habel U. The transsexual brain – a review of findings on the neural basis of transsexualism. *Neurosci Biobehav Rev* 2015; **59**: 251-66.
39. Money J, Simcoe K. Acrotomophilia, sex and disability: new concepts and case report. *Sex Disability* 1986; **7**: 43–50.
40. Bridy A. Confounding extremities: surgery at the medico-ethical limits of self-modification. *J Law Med Ethics* 2004; **32**: 148–58.
41. Brugger P. Der Wunsch nach Amputation. Bizarre Macke oder neurologische Störung? *Ars Medici* 2011; **2**: 59–63 (in German).

42. Mackenzie R, Cox S (2007) Transableism, disability and paternalism in public health ethics: taxonomies, identity disorders and persistent unexplained symptoms *Int J Law Context* 2007; **2**: 363–75.
43. McGeoch PD, Brang D, Song T, Lee RR, Huang M, Ramachandran VS. Xenomelia: a new right parietal lobe syndrome. *J Neurol Neurosurg Psychiatry* 2011; **82**: 1314–9.
44. Critchley M. The parietal lobes. London: Eward Arnold, 1953.
45. Brugger P, Lenggenhager B. The bodily self and its disorders: neurological, psychological and social aspects. *Curr Opin Neurol* 2014; **27**: 644–52.
46. Kasten E, Stirn A. Body integrity identity disorder (BIID): change of wish for amputation from left to right leg. *Z Psychiatrie Psychol Psychotherapie* 2009; **57**: 55-61.
47. Brang D, McGeoch, PD, Ramachandran VS. Apotemnophilia: a neurological disorder. *Neuroreport* 2008; **19**: 1305–6.
48. Romano D, Sedda A, Brugger P, Bottini G. Body ownership: when feeling and knowing diverge. *Consc Cog* 2015; **34**: 140–8.
49. Aoyama A, Krummenacher P, Palla A, Hilti LM, Brugger P. Impaired spatial-temporal integration of touch in body integrity identity disorder (BIID). *Spat Cog Comp* 2012; **12**: 96–110.
50. Romano D, Gandola M, Bottini G, Maravita A (2014) Arousal responses to noxious stimuli in somatoparaphrenia and anosognosia: clues to body awareness. *Brain* 2014; **137**: 1213–23.
51. Lenggenhager B, Hilti L, Brugger P. Disturbed body integrity and the "rubber foot illusion". *Neuropsychology* 2015; **29**: 205–11.
52. Van Stralen HE, van Zandvoort MJ, Kappelle LJ, Dijkerman HC. The rubber hand illusion in a patient with hand disownership. *Perception* 2013; **42**: 991–3.
53. Lopez C. The vestibular system: balancing more than just the body. *Curr Opin Neurol* 2016; **29**: 74–83.
54. Lenggenhager B, Hilti L, Palla A, Macauda G, Brugger P. Vestibular stimulation does not diminish the desire for amputation. *Cortex* 2014; **54**: 210–2.
55. Bottini G, Brugger P, Sedda A. Is the desire for amputation related to disturbed emotion processing? A multiple case study analysis in BIID. *Neurocase* 2015; **21**: 394–402.
56. Sterzi R, Bottini, G., Celani, M. G., et al. Hemianopia, hemianaesthesia, and hemiplegia after right and left hemisphere damage. A hemispheric difference. *J Neurol, Neurosurg Psychiatry* 1993; **56**: 308–10.
57. van Dijk M, van Wingen GA, van Lammeren A et al. Neural basis of limb ownership in individuals with body integrity identity disorder. *PLoS ONE* 2013; **8**, article e72212.
58. Karnath H-O, Baier B. Right insula for our sense of limb ownership and self-awareness of actions. *Brain Struct Funct* 2010; **214**: 411–7.
59. Gandola M, Invernizzi P, Sedda A et al. An anatomical account of somatoparaphrenia. *Cortex* 2012; **48**: 1165–78.
60. Ehrsson HH, Spence C, Passingham RE. That's my hand! Activity in premotor cortex reflects feeling of ownership of a limb. *Science* 2004; **305**: 875–7.
61. Hilti LM, Hänggi J, Vitacco DA et al. The desire for healthy limb amputation; structural brain correlates and clinical features of xenomelia. *Brain* 2013; **136**: 318–29.
62. Hänggi J, Bellwald D, Brugger P. Shape alterations of basal ganglia and thalamus in xenomelia. *NeuroImage: Clin* 2016; **11**: 760–69.
63. Jones CL, Ward J, Critchley HD. The neuropsychological impact of insular cortex lesions. *J Neurol Neurosurg Psychiatry* 2010; **81**: 611–8.
64. Gentile G, Bjornsdotter M, Petkova VI et al. Patterns of neural activity in the human ventral premotor cortex reflect a whole-body multisensory percept. *NeuroImage* 2015; **109**: 328–40.
65. Schilder P. (1935). The image and appearance of the human body. London: Kegan Paul.

66. Candini M, Aglioti SM. Visual and sensorimotor contributions to the esthetic appraisal of body form, motion, and emotion. *Europ Psychologist* 2015; **20**: 16–26.
67. Keysers C, Kaas JH, Gazzola V. Somatosensation in social perception. *Nat Rev Neurosci* 2010; **11**: 417–29.
68. Meltzoff AN. Infant imitation after a 1-week delay: long-term memory for novel acts and multiple stimuli. *Dev Psychol* 1988; **24**: 470–6.
69. Marshall PJ, Meltzoff AN. Body maps in the infant brain. *TICS* 2015; **19**: 499–505.
70. Bondeson J. A cabinet of medical curiosities. London: I.B. Tauris & Co, 1997.
71. Bolognini N, Rossetti A, Fusaro M, Vallar G, Miniussi C. Sharing social touch in the primary somatosensory cortex. *Curr Biol* 2014; **24**: 1513–7.
72. Banissy MJ, Ward J. Mirror-touch synaesthesia is linked with empathy. *Nat Neurosci* 2007; **10**: 815–6.
73. Goller A, Richards K, Novak S, Ward J. Mirror-touch synaesthesia in the phantom limb of amputees. *Cortex* 2013; **49**: 243–51.
74. Frank G. On embodiment: a case study of congenital limb deficiency in American culture. *Cult Med Psychiatry* 1986; **10**: 189–219.
75. Hilti LM, Brugger P. Incarnation and animation: physical versus representational deficits of body integrity. *Exp Brain Res* 2010; **204**: 315–26.
76. Smith RC. Body integrity identity disorder: a problem of perception? In: Alghrani A, Bennett R, Ost S, eds. Bioethics, medicine and the criminal law. Cambridge: Cambridge University Press, 2012, 71–87.
77. Ramachandran VS, Brang D, McGeoch P, Rosar W. Sexual and food preference in apotemnophilia and anorexia: Interactions between `beliefs' and `needs' regulated by two-way connections between body image and limbic structures. *Perception* 2009; **38**, 755–777.
78. Farah MJ. Why does the somatosensory homunculus have hands next to face and feet next to genitals? A hypothesis. *Neural Comp* 1998; **10**: 183–85.
79. Henderson WR, Smyth GE. Phantom limbs. *J Neurol Neurosurg Psychiatry* 1948; **11**: 88–112.
80. Grüsser SM, Winter C, Mühlnickel W et al. The relationship of perceptual phenomena and cortical reorganization in upper extremity amputees. *Neuroscience* 2001; **102**: 263–72.
81. Moulier V, Mouras H, Pelegrini-Issac M et al. Neuroanatomical correlates of penile erection evoked by photographic stimuli in human males. *NeuroImage* 2006; **33**: 689–99.
82. Taichman DB, Backus J, Baethge C et al. Sharing clinical trial data – a proposal from the international committee of medical journal editors. *N Engl J Med* 2016; **374**: 384–6.
83. Brugger P, Lenggenhager B, Giummarra MJ. Xenomelia: a social neuroscience view of altered bodily self-consciousness. *Front Psychol* 2013; **4**: article 204.
84. Elliott C. (2000). A new way to be mad. Atlantic Monthly, digital edition http://www.theatlantic.com/past/issues/2000/12/elliott.htm.
85. Baubet T, Gal B, Dendoncker-Viry S, Masquelet AC, Gatt MT, Moro MR. Apotemnophilia as a contemporary frame for psychological suffering. *Encéphale* 2007; **33**: 609–15 (in French).
86. Charland LC. A madness for identity: Psychiatric labels, consumer autonomy, and the perils of the internet. *Philos Psychiatry Psychol* 2005; **11**: 335–49.
87. Hacking I. Mad travelers: reflections on the reality of transient mental illnesses. London: University Press of Virginia, 1998.
88. Drescher J. Out of DSM: depathologizing homosexuality. *Behav Sci* 2015; **5**: 565–75.
89. BeauchampTL, Childress JF. Principles of biomedical ethics (6th Edition). Oxford: Oxford University Press, 2008.

90. Swindell JS. Two types of autonomy. *Am J Bioethics* 2009; **9**: 52–3.
91. White A. Body integrity identity disorder beyond amputation: consent and liberty. *HEC Forum* 2014; **26**: 225–36.
92. Barry PB. The Ethics of Voluntary Amputation. *Pub Affairs Quart* 2012; **26**: 1–18.
93. Johnston J, Elliott C. Healthy limb amputation: ethical and legal aspects. *Clin Med* 2002; **2**: 431–5
94. Müller S. Body integrity identity disorder (BIID) – is the amputation of healthy limbs ethically justified? *Am J Bioethics* 2009; **9**: 36–43.
95. Craimer A. The relevance of identity in responding to BIID and the misuse of causal explanation. *Am J Bioethics* 2009; **9**: 53–55.
96. Slatman J, Widdershoven G. Being whole after amputation. *Am J Bioethics* 2009; **9**: 48–49.
97. Kovacs J. Whose identity is it anyway? *Am J Bioethics* 2009; **9**: 44–45.
98. Kroeger K, Schnell T, Kasten E. Effects of psychotherapy on patients suffering from body integrity identity disorder. *Am J Applied Psychol* 2014; **3**: 110–5.
99. Patrone D. Disfigured anatomies and imperfect analogies: body integrity identity disorder and the supposed right to self-demanded amputation of healthy body parts. *J Med Ethics* 2009; **35**: 541–5
100. Dyer C. Surgeon amputated healthy legs. *Br Med J* 2000; **320**: 332.
101. Schwartz SJ, Lilienfeld SO, Meca A, Sauvigné KC. The role of neuroscience within psychology: a call for inclusiveness over exclusiveness. *Am Psychologist* 2016; **71**: 52–70.
102. Schlich T. Cutting the body to cure the mind. *Lancet Psychiatry* 2015; **2**: 390–2.

Figure Legends:

Figure 1. Functional brain correlates of xenomelia.
(A, B) Reduced responsivity of the right superior parietal lobe (arrows to blue outline on inflated surface models of two representative participants in A) to tactile stimulation of non-accepted parts of a leg in persons with xenomelia.[43] Bar graphs in B represent mean/standard deviation of the ratio between foot/thigh magnetoencephalographic activity of the superior parietal lobule (SPL) in 4 participants with xenomelia and 4 matched controls.
(C, D) Functional magnetic resonance imaging BOLD signal to tactile stimulation of the leg in 5 participants with an amputation desire for one leg.[57] (C) Red clusters indicate hyperactive regions in the participants with xenomelia compared to controls irrespective of the stimulated leg and comprise the right postcentral gyrus (PoCG), bilateral precentral gyrus (PrCG), right premotor cortex (PMC) and the right insula (Ins). (D) Blue clusters represent hypoactive premotor cortex (PMC) in response to tactile stimulation (affected vs. unaffected leg in xenomelia compared with the corresponding limbs of the controls). Color bars represent the error probability. Panels A and B adapted from ref. 43; courtesy of David Brang and Paul McGeoch; panels C and D reproduced, with permission, from Figs. 1 and 2 of ref. 57.

Figure 2. Structural brain correlates of xenomelia.
(A) Surface-based morphometry in participants with a desire for leg amputation and matched controls (n=13, each).[61] Regions of reduced cortical thickness and surface area, respectively, comprised the right superior parietal lobule (SPL), the right inferior parietal lobule (IPL), primary (SI) and secondary somatosensory cortex (SII) as well as the right anterior insula (AIC) (left panel). The strength of an individual's amputation desire as rated on the Zurich

Xenomelia Scale (ZXS)[49] was inversely related to the cortical surface area within the right IPL cluster (right panel).
(B-D) Negative and positive tissue displacements revealed by shape analyses of subcortical structures in the same participant population.[62] (B) Local thinning (blue clusters) in xenomelia is evident in the left dorsomedial putamen (Put) and left ventral caudate nucleus (CN). (C) Local thickening (blue clusters) is evident mainly in the right lateral pallidum. (D) Thickening of the left anterior lateral thalamus (blue clusters). Tissue displacements in all these subcortical structures are mainly localized in subregions housing a somatotopic representation of the extremities or projecting to sensorimotor cortical regions.
The color bar represents error probability in (A) and the statistical values of the multivariate shape analysis in B–D. The direction of the effects (thinning or thickening) is coded in the displacement vectors (small arrows; invisible in panel B due to inward direction). Panel A reproduced with permission from Oxford University Press, panels B-D with permission from Elsevier.

Panel 1:

Illustrative first-hand descriptions of the desire for…

(a) limb amputation:

"*I feel myself complete without my left leg … I'm over-complete with it.*"[1, p. 922]

"*I started out as a devotee but the need to see and be with an amputee became so strong that I knew the answer to my problem was to have my own stump.*" [2,p.16]

"*The soul feels as though it belongs to a body with only one leg. The body does not correspond to this inner reality.*" [3,p.17]

"*Since my 13th year, my conscious life has been absorbed, with varying intensity, in a bizarre and prepotent obsessive wish, need, desire to have my leg amputated above the knee*." [4,p.117]

"*I want to wake up with a bandaged stump with two crutches by my bed which I wish to depend on for the rest of my life.*" [5,p.85]

"*I was 4 years old when I first knew that I wanted my leg cut off. I have no idea how I got this 'need' or where it came from.*" [6,p.103]

(b) paraplegia:

"*I have needed to be paralysed from the belly-button downwards since I was a very young child. […] The only thing that has helped me feeling some peace of mind is to use a wheelchair. In my mid to late 20's, I started living 'full-time' - That is, I used a wheelchair in my day-to-day life, all day, every day*." [7,p.88]

"*I am using a wheelchair 'full-time' when I'm in public. I walk at home. This is the only way how to remain somewhat functional*."[8,p.3]

(c) orchiectomy:

"*[My] testicles seemed unnatural; a growth that shouldn't be there.*" [9,p.854]

(d) sensory impairment:

"*I want to be deaf but I'd also use hearing aids to restore my hearing"*. [10,p.190]

"*When it 's pitch dark, I come closest to life as it should be for me"*. (own observation, unpublished)

Panel 2

Characteristic features of disability desires and the persons affected

Established:
- marked suffering
- mostly men affected
- typically high education status
- largely unremarkable personality profile
- first manifestation in childhood or early adolescence
- "pretending" the desired disability almost universally present
- most common form is the desire for limb amputation (xenomelia)
  - more often for leg(s) than arm(s)
  - more often for left-sided than right-sided limbs

Controversial:		
emasculation desire part of BIID?	yes [9,12]	no [13]
paraphilic component required?	yes [14,15]	no [1,16]
association with non-heterosexual orientation?	yes [8,17]	no [18,19]
key experience as a trigger?	rather yes [1,19]	rather no [16]
psychotherapy a successful treatment option?	yes [20]	no [21]
surgery a successful treatment option?	yes [1,8,21]	no [22]

Panel 3

Proposed diagnostic criteria for disability desires as "body integrity identity disorder" (BIID)

A An intense and persistent desire to become physically disabled in a significant way (e.g., major limb amputee, paraplegic, blind), with onset by early adolescence

B Persistent discomfort or intense feelings of inappropriateness concerning current nondisabled body configuration

C The desire to become physically disabled results in harmful consequences, as manifested by either (or both) of the following:
  (1) The preoccupation with the desire (including time spent pretending to be disabled) significantly interferes with productivity, with leisure activities, or with social functioning (e.g., person is unwilling to have a close relationship because it would make it difficult to pretend)
  (2) Attempts to actually become disabled have resulted in the person putting his or her health or life in significant jeopardy

D *The desire to become disabled is not primarily motivated by sexual arousal* or by any perceived advantages of becoming disabled

E The disturbance is not a manifestation of a psychotic process (e.g., desire to amputate a limb because of delusional conviction that the limb belongs to another person), is not due to a primary neurological condition such as post stroke neglect syndrome and is not better accounted for by another mental disorder such as Body Dysmorphic Disorder or Factitious Disorder

Subtype based on predominant desired disability
  Amputation type
  Paraplegia type
  Other type
  Unspecified type

Reprinted, with permission, from First and Fisher, 2012 (ref. 16, Table 1); first part of point D (italics ours) problematic, according to the present review (see text).

Panel 4

Future directions

Empirical research…	… should expand to include variants of disability desires different from xenomelia (i.e. include persons who desire sensory impairments etc.)
	… should expand to include persons with gender dysphoria, anorexia and related disturbances of bodily experience
	… should move on from investigations of an individual's body schema to probing higher-order processes of body representation that link a person to society (e.g., mimicry, sexuality, empathy)
	… should plan crosscultural studies
	… should consider data sharing in the case of neuroimaging investigations akin to requirements proposed for clinical trials (ref. 82)
Nosology…	… should consider labelling disability desires "body dysphoria" (BD) and further examine the conceptual vicinity to gender dysphoria (GD) and the bodily dysphorias associated with eating disorders

Table 1

Survey	**Sample characteristics**		**Ratios in %**			
Reference (chrono-logical order); type of inquery	**Size; gender; age distribution**[a]	**Age at onset**[a]	**legs:arms**	**left:right:bilateral**	**sexual orien-tation hetero:homo: other**[b]	**handedness right:non-right**
First, 2005[1] ; telephone interviews	n=52 (4 women, 1 intersex); mean age 48.6 (range 23-77)	in 65% before age 8, in 94% before age 16 (mean n.r.)	76:24 [c]	55:27:18 [d]	61:31:7	n.r.
Blanke et al., 2009[18]; telephone interviews	n=20 (3 women); mean age 48.4 (range 29-72);	in 65% between ages 3 and 9 (mean = 11.6)	80:20	35:20:45	90:10:0	90:10
Kasten, 2009 [3]; postal survey using standardi-zed personality inventories	n=9 men[e] from early 30s to early 70s	in 67% at or before age 8 (mean = 8), range 4 - 12	100:0	60:20:20	33:56:11	n.r.
Johnson et al., 2011[**17**]; internet questionnaire [f]	n=72 (8 women, 3 "other"), mean age 46 (SD=16)	n.r.	81:10 (n=8 arm-leg-combinations)	42:28:30[g]	60:25:14	78:22
Blom et al., 2011[8]; internet-administered standardized psychiatric inventories	n=54[h] (79.6% men); age range 18-76	mean = 6.7, range 3 - 15	90:7 (n=1 with desire for tetramelia)	37:30:33[i]	56:28:17[i]	73:27[i]
Noll and Kasten, 2014[21]; internet questionnaire	n=18[j] (3 women), age range 27-73; all have achieved the desired amputation	n.r.	86:6 (n=1 arm-leg-combination)	50:22:28	76:10:14	n.r.

Oddo et al., 2014[23]; individual examination, standardized persona-lity inventories	n=15 men[k] mean age 50 (range 32-68)	n.r.	100:0	n.r. (80% unilateral)	40:40	n.r.

Table 1: Characteristics of the desire for amputation in seven questionnaire studies

bilat. = bilateral; n.a. = not applicable; n.r. = not reported;

[a] age in years
[b] other = bisexual or asexual
[c] 100% = those 50 individuals wishing for a major limb amputation
[d] 100% = those 44 individuals, who specified laterality
[e] 1 man with desire for paraplegia
[f] describes two surveys (total n=97, but some individuals responded to both); numbers here refer to larger sample of survey 2
[g] 100% = all cases with an amputation desire
[h] includes n=2 with desire for blindness and n=2 with desires for other sorts of physical disability
[i] 100% = 30 individuals with a limb amputation desire
[j] three participants with non-amputation desires not considered here
[k] data of 1 woman not analyzed, data of 2 more men also not analyzed (1 amputated, 1 with insufficient data)
[l] 20% n.r.

Table 2:

Compared feature	**Disability Desires**	**Gender Dysphoria**
Marked distress due to body morphology	yes	yes
Typical age of onset	early	early in FtM, both early and late in MtF
Sex ratio (biological)	male >female	male >female
Elevated prevalence of non-right-handedness	yes	yes[13]
Simulation of desired state	frequent (“pretending”)	frequent (“crossdressing”)
Sexual arousal associated with simulation	frequent	frequent in gynephilic MtF[14]
Psychotherapy an effective treatment?	no	no
Surgery an effective treatment?	yes (illegally available in some non-Western countries)	yes (legally available in many countries)
Secondary psychiatric disorders	frequent	frequent
Co-occurrence described?	yes[1,16]	

Table 2: Similarities between disability desires and gender dysphoria (MtF = male-to-female transsexuals, FtM = female-to-male transsexuals)

Table 3

Value orientation (following Beauchamp & Childress)[89]	**Arguments in favor of elective disability**	**Arguments against elective disability**
Respect for autonomy: One should respect the decision-making capacities of autonomous persons; one should enable individuals to make reasoned informed choices. ***Major issue:*** *Are the conditions for respecting the autonomy of the patient met?*	Supporters claim that the conditions for respecting the autonomy of the patients are fulfilled in at least some patients: - The decision is informed and the patients have a reasonable understanding on what it is like to have the desired condition (expressed, beside others, by pretending) - The patients have decision-making capacity; it's not the decision that needs to be rational but the rationality of the thought process that cumulates in the decision. - The degree of pressure from non-rational considerations ("looping effect") is comparable to pressure (advertising, gender-norm) in other types of interventions like plastic surgery where autonomy is granted to the patient. - The choice is among the medically reasonable alternatives (see below). Patients who fulfill these conditions can rely on the principle of respecting autonomy; this would be the case particularly for certain cases of limb amputation.[76,90,91]	Detractors agree that patients can refer on the principle of respecting autonomy if the conditions on the left side are met – but they question fulfillment of the conditions: - The patients lack crucial knowledge on how it would be to have the desired disability and thus do not provide consent that can be considered "informed". - Having an irrational wish puts into question that the patients have decision making capacity with respect to this desire. - There are indications that a disability desire is a psychiatric disease 'manufactured' in cyberspace - There are alternatives (psychotherapy, medication) which are much less invasive; furthermore, research should be advanced instead of allowing an irreversible intervention. Furthermore, even if the principle of respecting autonomy is granted, the main aim of this principle is to protect patients from unwanted interventions – but it does not justify to request an intervention – in particular in cases where the surgeon would face criminal liability.[92-94]

Nonmaleficence: One should avoid causing harm in the patient (but also with respect to other involved persons); the harm should not be disproportionate to the benefits of treatment. ***Major issue:*** *How severe is the harm caused by disability?*	Supporters weight the (psychological) harm of suffering from a disability desire higher than the (physiological) harm of having a disability. They also claim that physical disability need not result in significantly lower life satisfaction, even for people who did not choose to become disabled. Finally, elective disability will prevent sufferers from taking matters into their own hands, with potentially disastrous consequences; in particular in case of amputations.[33,95]	Detractors consider disabilities as paradigmatic cases of harm – in particular in case of sensory disability (blindness, deafness). For example, amputations bear great risks and often have severe consequences besides the disability, for example, infections, thromboses, paralyses, necrosis, or phantom pain. They also remark that medical history is filled with surgical treatments for psychiatric problems; many of those today appear to have been seriously misguided. Only in extreme cases with high risk of self-injury, elective amputations might be an option.[93,94]
Beneficence: One should act for the benefit of the patient; one should take positive steps to prevent and to remove harm from the patient. ***Major issue:*** *How credible are reports that elective disabilities have increased the well-being of patients?*	Supporters interpret the desire for a disability in some patients as a set of stable values that are crucial for the person; to approve of the person's decision is to respect his or her bodily integrity. For the case of amputation they refer to the case of patients with amputations due to ischemia who gain enormous relief from their symptoms – an amputation in a patient suffering from a disability desire is seen as equally relevant for the wellbeing of this person.[76,96]	Detractors question the scientific evidence gained so far with respect to the effectiveness of amputations for the wellbeing of patients. The number of cases is too low and one could expect mechanisms of self-deception in those rare cases: after having invested enormous emotional resources in getting a procedure that is not only irreversible, but which they have always seen as the only possible solution to their problems, some may find it difficult to admit to themselves that it has been a mistake. [93,94]
Justice: Patients in similar positions should be treated in a similar manner (individual level); benefits, risks and costs should be distributed fairly	With respect to the individual level, supporters see an analogy to sex-reassignment surgery that involves irreversibly losing one's capacity for reproduction – and there it has been accepted that surgery is appropriate for treating Gender Identity Disorder.	Detractors consider analogies of disability desires with other examples where demanded (or refused) interventions lead to body modifications or even harm as not sufficiently supported. They furthermore weight societal costs (medical treatment, rehabilitation, early

(societal level). ***Major issue:*** *How plausible are analogies with respect to fairness on the individual and societal level?*	With respect to the societal level, they point to reports that those who achieve amputation become more productive, happy and contributing members of the community and do no longer consume psychiatric resources. This counteracts the resources needed for performing interventions and support requirements that a functional handicap involves. [76,97]	retirement, etc.) higher. Generally, they claim that the empirical data for assessing the potential economic benefit of elective disability is poor and unlikely to be positive. Justice considerations with respect to resource allocation should rather focus on research on the causes of disability desires for finding alternative, less invasive treatment options. [94,98]

Table 3: Pros and cons in the ethical debate about elective disability